CASE REPORT

Coronary involvement in the Churg-Strauss syndrome

S Hellemans, J Dens, D Knockaert

Abstract

The Churg-Strauss syndrome (CSS) is a rare systemic disease characterised by vasculitis and peripheral eosinophilia in patients with an atopic constitution. Cardiac involvement is an important cause of morbidity and mortality yet coronary involvement is very rarely documented. We report the case of a 38 year old man presenting with fulminant heart failure. Coronary arteriography demonstrated extensive focal vasculopathy consistent with vasculitis. The diagnosis of CSS was established based upon the classical diagnostic criteria and corticosteroid treatment resulted in a spectacular remission of disease activity.

(*Heart* 1997;77:576–578)

Keywords: Churg-Strauss syndrome; coronary vasculitis

Department of Cardiology, Gasthuisberg University Hospital, Catholic University Leuven, Leuven, Belgium
S Hellemans
J Dens

Department of General Internal Medicine
D Knockaert

Correspondence to:
Dr Hellemans, Department of Cardiology, Gasthuisberg University Hospital, Herestraat 49 B3000 Leuven, Belgium.

Accepted for publication 27 March 1997

The Churg-Strauss syndrome (CSS) or allergic angiitis and granulomatosis is a rare primary systemic vasculitis, typically characterised by a history of asthma or allergy, and by the occurrence of eosinophilia.[1] Seventy seven cases were examined at the Mayo Clinic during 1950–92 and a recent epidemiological study estimated the annual incidence of CSS to be 2·4 per million population.[2 3] Cardiac involvement was shown in up to 60% of the post-mortem examinations in the original report of Churg and Strauss, and accounted for 50% of deaths in another series.[1 4 5] Cardiac involvement is mostly thought to result from an interstitial myopathic process but vasculitis of the coronary vessels is documented extremely rarely.[4–6] We describe a typical case of CSS presenting as fulminant cardiac failure with angiographically documented coronary vasculitis.

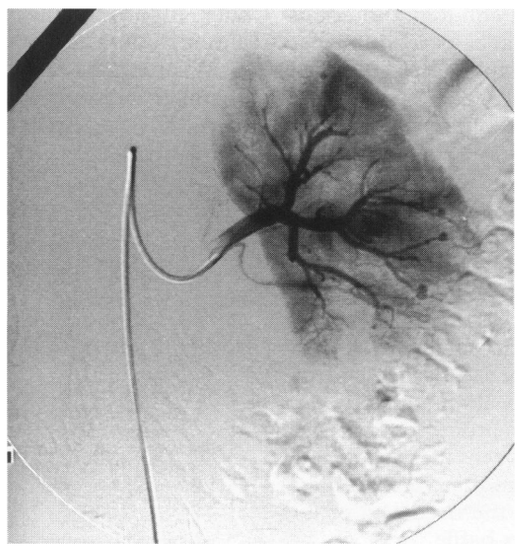

Figure 1 Renal arteriogram revealing multiple microaneurysms at the distal branches of the left renal artery.

Case report

A 38 year old man with a five year history of asthma was admitted to another hospital in November 1995 because of severe vomiting and diarrhoea. The peripheral blood eosinophil count was 48% at that time. No underlying disease could be detected and he improved spontaneously. One month later he was readmitted because of fever. Despite extensive investigation no infectious cause could be found and five days later diffuse pulmonary infiltrates developed. Echocardiography revealed left ventricular failure. Because of respiratory distress, mechanical ventilation was needed, and inotropic support with dobutamine was instituted. At that time, he was transferred to our hospital with the diagnosis of cardiorespiratory failure due to myocarditis. The referring cardiologist considered the patient as a possible candidate for cardiac transplantation.

On admission, his temperature was 37°C and white blood cell count was 25·5 × 10^9/l with a shift to the left but no eosinophilia. Urine analysis revealed proteinuria, the sediment was normal. The electrocardiogram showed sinus tachycardia, incomplete right bundle branch block, and diffuse abnormal repolarisations.

On echocardiography, the left ventricular ejection fraction was 39%, the right ventricular function was normal as were the heart valves, and no pericardial effusion was present.

Because of suspicion of vasculitis, treatment with corticosteroids (prednisone 1 mg/kg) was started after performing bronchoscopy with bronchoalveolar lavage which was normal. Endomyocardial biopsy documented noduli with a central amorf necrosis surrounded by histiocytes resembling Asschoff bodies.

The patient was weaned from the ventilator after 72 hours. Electromyography revealed sensorimotory polyneuropathy, and renal

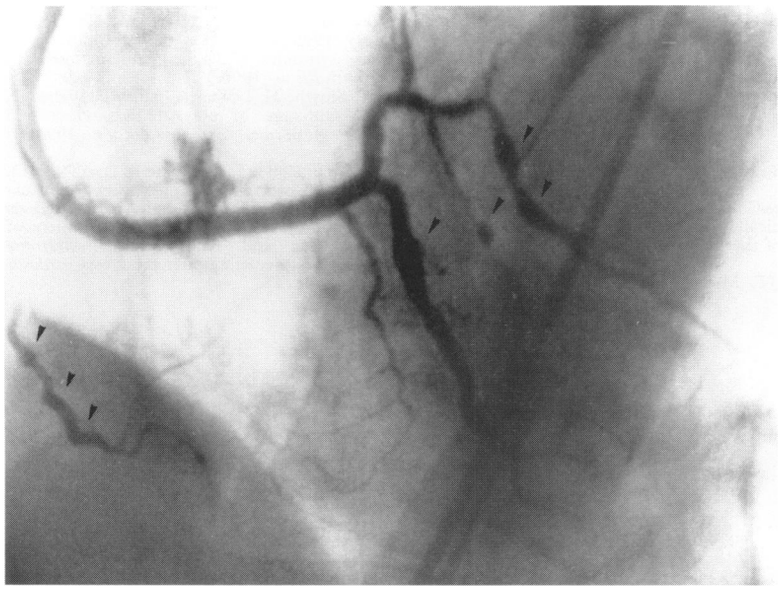

Figure 2 Coronary arteriogram showing vessel wall irregularities, abrupt terminations, and small aneurysms at the posterior descendens and two inferolateral branches of the right coronary artery.

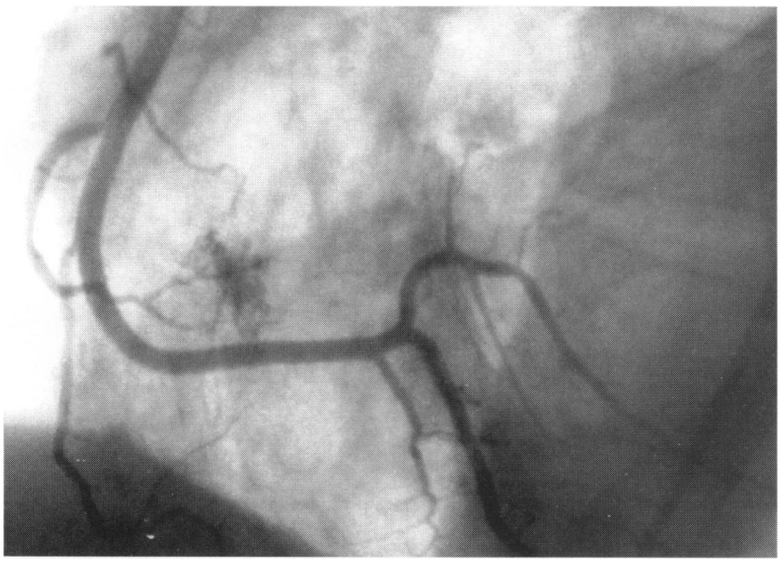

Figure 3 Coronary arteriogram after six months of corticosteroid treatment showing important regression of the focal vasculopathy.

angiography, performed because of important proteinuria, revealed multiple microaneurysms (fig 1). Coronary arteriography was performed one week later and showed extensive focal vasculopathy with multiple vessel wall irregularities, abrupt terminations, and small aneurysms at a diagonal branch of the left anterior descending coronary artery, the posterior descendens, and two inferolateral branches of the right coronary artery (fig 2).

Six months later the patient was asymptomatic on treatment of 6 mg prednisone once a day. A repeat coronary arteriography showed spectacular regression of the lesions (fig 3), and ventriculography showed normal left ventricular function.

Discussion

The patient presented with angiographically proven vasculitis of the coronary and kidney arteries. The multiple vessel wall irregularities, multiple abrupt terminations, and the small aneurysms (fig 2) were diagnostic for vasculitis. This can also be found in rheumatoid arthritis, polyarteritis nodosa, Kawasaki disease, and systemic lupus erythematosus. Because of the history of asthma, the documented eosinophilia of 48% on differential white blood cell count, the presence of pulmonary infiltrates, and polyneuropathy, this case met both sets of criteria for the diagnosis of CSS, recently developed by the American College of Rheumatology.[7] CSS typically has three phases and this was also the case in our patient. The initial prodromal phase consists of allergic rhinitis, nasal polyposis or bronchial asthma. This is followed by a period of peripheral and tissue eosinophilia commonly associated with pulmonary infiltrates. The third phase is characterised by a systemic vasculitis that can be fulminant and life threatening.[5] This vasculopathy preferentially affects the lungs, skin, heart, and peripheral nerves. Renal involvement is less frequent but was present in our case. Indeed the proteinuria suggested glomerulonephritis but, because of suspicion of vasculitis, we performed renal angiography before considering kidney biopsy. The presence of multiple microaneurysms was considered a contraindication for biopsy.

The cardiac involvement in CSS consists of pericarditis and occasionally tamponade, myocarditis, which can lead to terminal restrictive or congestive heart disease, and myocardial infarction.[4-6] Endomyocardial biopsy rarely shows acute inflammatory changes and the presence of eosinophils, necrotising vasculitis or extravascular granuloma is often non-diagnostic. To evaluate the cause of myocardial infarction, chest pain or heart failure in patients with CSS, coronary arteriography has been performed in some cases and usually no abnormalities were found. Coronary vasculitis was found at necropsy in six of 10 cases originally reported by Churg and Strauss.[1 6] Coronary involvement has rarely been found premortem and to our knowledge this case represents the first abnormal coronary arteriogram reported in a patient with CSS who survived. Another case with an abnormal coronary arteriogram was recently reported but this patient died.[6] Cardiologists are not familiar with vasculitis syndromes because major cardiac problems are rarely presenting manifestations. This case underlines the role of an aggressive invasive diagnostic approach in patients with evidence of cardiac involvement by a vasculitis syndrome. It also demonstrates the possible dramatic reversal of severe cardiac disease with steroid therapy.[8 9] In our patient no additional immunosuppressive therapy was required. Delay in recognising the cardiac involvement probably accounts for the important cardiac mortality in CSS.[1 6 8]

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